WE CLAIM:

1. A human recombinant lysosomal enzyme or variant thereof produced in END3 complementation group CHO cells, or a derivative of said enzyme or variant, wherein said enzyme has a high level of phosphorylation and a low level of unphosphorylated high-mannose oligosaccharides.

2. The enzyme of claim 1, wherein said enzyme is selected from the group consisting of: acid alpha glucosidase, aspartylglucosaminidase, acid lipase, cysteine transporter, Lamp-2, α-galactosidase A, acid ceramidase, α-L-fucosidase, β-hexosaminidase A, GM2-activator deficiency, α-D-mannosidase, β-D-mannosidase, arylsulfatase A, saposin B, neuraminidase, α-N-acetylglucosaminidase phosphotransferase, phosphotransferase γ-subunit, L-iduronidase, iduronate-2-sulfatase, heparan-N-sulfatase, α-N-acetylglucosaminidase, acetylCoA:N-acetyltransferase, N-acetylglucosamine 6-sulfatase, galactose 6-sulfatase, β-galactosidase , N-acetylgalactosamine 4-sulfatase, hyaluronoglucosaminidase, multiple sulfatases, palmitoyl protein thioesterase, tripeptidyl peptidase I, acid sphingomyelinase, cholesterol trafficking, cathepsin K, α-galactosidase B, and sialic acid transporter.

3. A human recombinant acid alpha glucosidase (rhGAA), or variant thereof produced by END3 complementation group CHO cells, or a derivative of said enzyme or variant, wherein said rhGAA has a high level of phosphorylation and low level of unphosphorylated high-mannose oligosaccharides.

4. The enzyme of any one of claims 1-3, wherein the END3 complementation group CHO cell is a G71 cell line or derivative thereof.

5. A method for producing highly phosphorylated human recombinant lysosomal enzymes or variants thereof, comprising the steps of:

(a) culturing Chinese Hamster Ovary (CHO)-derived END3 complementation group cells;

(b) preparation of a mammalian expression vector suitable for said END3 complementation group cells;

(c) transfection of said END3 complementation group cells with said expression vector;

(d) selection and cloning of a END3 complementation group transfectant; and

(e) optimization of cell culture process methods for manufacturing.

6. The method of claim 5, wherein said enzymes have a low level of unphosphorylated high-mannose oligosaccharides.

7. A lysosomal enzyme, variant or derivative thereof produced by the method of claim 5.

8. A composition comprising the lysosomal enzyme, variant or derivative of claim 7 and a pharmaceutically acceptable carrier, diluent or excipient.

9. The method of any one of claims 5-6 wherein the END3 complementation group CHO cell is a G71 cell line or derivative thereof.

10. A method for producing highly phosphorylated human recombinant acid alpha glucosidase (hrGAA) or variant thereof, comprising the steps of:

(a) culturing Chinese Hamster Ovary (CHO)-derived END3 complementation group cells;

(b) preparation of a mammalian expression vector suitable for said END3 complementation group cells;

(c) transfection of said END3 complementation group cells with said expression vector;

(d) selection and cloning of a END3 complementation group transfectant; and

(e) optimization of cell culture process methods for manufacturing.

11. The method of claim 10, wherein said hrGAA has a low level of unphosphorylated high-mannose oligosaccharides.

12. A highly phosphorylated recombinant acid alpha glucosidase (hrGAA), variant or derivative thereof produced by the method of claim 10.

13. A composition comprising the recombinant acid alpha glucosidase, (hrGAA), variant or derivative thereof of claim 12 and a pharmaceutically acceptable carrier, diluent or excipient.

14. The method of any one of claims 10-11 wherein the END3 complementation group CHO cell is a G71 cell line or derivative thereof.

15. A method of treating a deficiency of a lysosomal enzyme comprising administering to a subject in need of said lysosomal enzyme, a therapeutically effective amount of said lysosomal enzyme, wherein said lysosomal enzyme is a human recombinant lysosomal enzyme, or variant thereof produced by CHO-derived END3 complementation group cells, or a derivative of said enzyme or variant.

16. The method of claim 15, wherein said lysosomal enzyme deficiency is selected from the group consisting of: aspartylglucosaminuria, cholesterol ester storage disease, Wolman disease, cystinosis, Danon disease, Fabry disease, Farber lipogranulomatosis, Farber disease, fucosidosis, galactosialidosis types I/II, Gaucher disease types I/II/III, Gaucher disease, globoid cell leukodystrophy, Krabbe disease, glycogen storage disease II, Pompe disease, GM1-gangliosidosis types I/II/III, GM2-gangliosidosis type I, Tay Sachs disease, GM2-gangliosidosis type II, Sandhoff

disease, GM2-gangliosidosis, α-mannosidosis types I/II, β-mannosidosis, metachromatic leukodystrophy, mucolipidosis type I, sialidosis types I/II mucolipidosis types II /III I-cell disease, mucolipidosis type IIIC pseudo-Hurler polydystrophy, mucopolysaccharidosis type I, mucopolysaccharidosis type II, Hunter syndrome, mucopolysaccharidosis type IIIA, Sanfilippo syndrome, mucopolysaccharidosis type IIIB, mucopolysaccharidosis type IIIC, mucopolysaccharidosis type IIID, mucopolysaccharidosis type IVA, Morquio syndrome, of mucopolysaccharidosis type IVB Morquio syndrome, mucopolysaccharidosis type VI, mucopolysaccharidosis type VII, Sly syndrome, mucopolysaccharidosis type IX, multiple sulfatase deficiency, neuronal ceroid lipofuscinosis, CLN1 Batten disease, Niemann-Pick disease types A/B, Niemann-Pick disease, Niemann-Pick disease type C1, Niemann-Pick disease type C2, pycnodysostosis, Schindler disease types I/II, Schindler disease, and sialic acid storage disease.

17. The method of any one of claims 15-16 wherein the END3 complementation group CHO cell is a G71 cell line or derivative thereof.